Adams-Stokes syndrome caused by sinoatrial block

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Forty-six patients with syncope and/or black-out episodes due to sinoatrial block are presented. Male-to-female ratio was 1 to 1, and mean age at onset of symptoms approximately 63 years in both sexes. Additional heart disease, especially coronary artery disease, was frequently, but not constantly, found. Twenty-five per cent had sinus node dysfunction as the only manifestation of cardiac disease. Ninety per cent had frequent paroxysms of one or several types of a wide spectrum of supraventricular dysrhythmias, which often gave rise to troublesome symptoms in the form of palpitation, fatigue, and/or congestion, but also made it easier to recognize the cardiac origin of the unspecific cerebral symptoms. Cerebral symptoms occurred suddenly and unexpectedly, and often disabled the patients to a severe degree. Drug treatment proved of little value, but pacemaker treatment stopped the cerebral attacks in all the 39 cases in which it was performed. In 3 out of 24 patients pacemaker implantation also abolished the tendency for paroxysms of tachydysrhythmias; in the remaining 21 it allowed for maximal drug treatment which led to nearly total relief from tachydysrhythmias in 17.

Attacks of third-degree sinoatrial block, resulting in cerebral ischaemic symptoms, account for 10 to 20 per cent of all cases, in which artificial pacemakers are considered necessary (Rasmussen, 1971; Jensen *et al.*, 1973). Syncope and black-out episodes have correspondingly been recognized as important features of the so-called sick sinus syndrome, caused by a wide spectrum of supraventricular dysrhythmias with often alternating appearance in the same individual (Ferrer, 1968; Slama *et al.*, 1969; Eraut and Shaw, 1971; Rasmussen, 1971; Mandel *et al.*, 1972; Rubinstein *et al.*, 1972).

The aim of the present paper is to focus on problems of diagnosis, aetiology, electrophysiology, and clinical management of this special type of Adams-Stokes disease. The paper is based on a consecutive series of 46 patients who had all experienced syncope/or black-out episodes due to paroxysmal third-degree sinoatrial block.

Electrocardiographic tracings usually do not permit any differentiation between third-degree sinoatrial block, sinus arrest, and atrial standstill (Schamroth, 1971). In this paper the term 'third-degree sinoatrial block' has been used as a common designation for electrocardiographic abnormalities consistent with either one of the mentioned conditions.

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Patients

There were 46 patients with a chronic, intermittent tendency to syncope and/or black-out episodes, assumed to be caused by paroxysms of third-degree sinoatrial block. In this context 'syncope' is defined as sudden complete loss of consciousness and 'black-out' episodes as momentary mental clouding and loss of vision without complete unconsciousness. The series includes all patients with this condition seen at medical department B, Rigshospitalet, Copenhagen, from 1965 to 1972.

In 27 cases, electrocardiograms immediately before and during the fainting showed prolonged periods of third-degree sinoatrial block and ventricular asystole, whereas no electrocardiograms in relation to cerebral symptoms were obtained in the remaining 19 cases. In the latter cases a presumptive diagnosis of cardiac syncope caused by paroxysmal third-degree sinoatrial block was made probable by electrocardiograms which showed the following.

- Paroxysms of third-degree sinoatrial block and ventricular asystole in 8 patients. The paroxysms lasted for as long as 3 to 15 seconds, but were nevertheless unaccompanied by cerebral symptoms.
- Frequent changes between sinus rhythm (most often sinus bradycardia) and atrioventricular junctional rhythm and/or second-degree sinoatrial block in 11 patients.

The diagnoses were finally supported by the outcome of pacemaker implantation which was performed in 39 patients and in all cases stopped the cerebral attacks.

Daily 12-lead electrocardiograms were recorded in all

patients, and there was an average of 6.5 days of continuous electrocardiographic monitoring (range 2 to 21 days).

Patients in whom the sinus node dysfunction might be attributed to drug intoxication, acute myocardial infarction, electrolyte disturbances, or myxoedema are not included in the series.

Results

The series included 22 men and 24 women. Age range at onset of cerebral symptoms was 20 to 90 years. Mean age for the series as a whole was 63.4 years (SD 12.4 years), 65.2 for men and 61.6 for women. The difference in number and age between the two sexes was not statistically significant (Fig. 1).

Functional capacity class (New York Heart Association Criteria Committee) and subjective symptoms between attacks are shown in Table 1.

Five patients had syncopal attacks lasting more than 30 minutes. Thirty-one patients had attacks lasting a few minutes, and of these 19 also had black-outs. Ten patients had black-outs only. Eleven patients had prodromal symptoms: five experienced slow or fast heart rate immediately before attacks, 5 had more vague feelings of malaise, and one had anginal pain and dyspnoea immediately before attacks.

Twenty-four patients had had more than 10 syncopal attacks for more than a year before the diagnosis and altogether 29 had had more than 10 attacks (Table 2).

Additional heart disease, heart size, and presence of congestive heart failure appear in Fig. 2. Seven patients had been operated on for gallbladder disease, and another 5 had recurrent gallbladder symptoms. Two patients had diabetes mellitus. Four patients had had a febrile illness within 3 months before the onset of the cerebral attacks. None had

TABLE 1 Cardiac symptoms and functional capacity class, according to criteria of New York Heart Association

	No. of patients
Functional capacity	
class I	17 (37%)
class II	20 (43%)
class III-IV	9 (20%)
Subjective symptoms	
anginal pain	12 (26%)
shortness of breath at rest	9 (20%)
shortness of breath at exertion	13 (28%)
peripheral oedema	9 (20%)
sensation of rapid heart action	22 (48%)
sensation of slow heart action	6 (13%)
fatigue, malaise	32 (70%)
no heart symptoms	3 (7%)

FIG. 1 Sex and age.

TABLE 2 Number of cerebral ischaemic episodes and observation time

No. of episodes	Observation time (days)				Total no. of patients
	< 7	7-30	31-365	> 365	
1-2	0	3	2	1	6
3-10	0	1	4	6	11
> 10	2	3	7	17	29
Total no. of patients	2	7	13	24	46

myxoedema. Six patients had been treated with digitalis: in all cases digitalis was discontinued without changes in the frequency or severity of the syncopal attacks.

Between cerebral symptoms, periods of atrial fibrillation, flutter, or tachycardia were seen in 24 patients (Table 3), and 37 patients had periods of sinus bradycardia, second-degree sinoatrial block, or junctional rhythm (Table 4). Twenty patients had alternating periods of tachy- and bradydysrhythmia between attacks, and 5 patients had consistently normal sinus rhythm (Fig. 3). Other transient electrocardiographic abnormalities noted were atrial escape beats in 3 patients, wandering pacemaker in 3 patients, left atrial rhythm (negative P waves in all standard leads and in lead V6) in 1 patient, and reciprocal rhythm of atrial origin in 4 patients. Two patients had Wenckebach periodicity simultaneously in sinoatrial and atrioventricular conduction, characterized by a cyclic grouping of beats, each cycle of 3 to 4 beats having progressive diminution of the PP intervals and simultaneous progressive

FIG. 2 Additional heart disease, heart size, and congestive heart failure.

TABLE 3 Observed types of tachydysrhythmias

	Total	No. of patients		
		One type of tachydysrhythmia only	With two or several tachydysrhythmias	With tachydysrhythmia as predominant finding
Atrial tachycardia	7 (15%)	5	2	1
Atrial flutter	11 (24%)	1	10	1
Atrial fibrillation	18 (39%)	7	11	3
No tachydysrhythmia observed	22 (48%)	—	—	—

FIG. 3 Incidence of tachy- and bradydysrhythmias (episodes of third-degree sinoatrial block not included).

prolongation of the PR intervals and terminated by a dropped P-QRS complex. Two patients had, during junctional escape rhythm, Wenckebach periodicity in a retrograde atrial conduction, i.e. progressive prolongation of the RP' interval with occasional dropped P' waves. Three patients had Wenckebach exit block in junctional escape rhythm, diagnosed by a progressive diminution of the RR intervals followed by a long RR interval.

In 23 patients episodes of prolonged ventricular asystole due to third-degree sinoatrial block were

TABLE 4 Observed types of bradydysrhythmias (episodes of third-degree sinoatrial block not included)

	Total	No. of patients		
		One type of bradydysrhythmia only	With two or several bradydysrhythmias	With bradydysrhythmia as predominant finding
Sinus bradycardia (or constant second-degree SA block)	21 (45%)	2	19	3
Second-degree SA block without Wenckebach phenomenon	19 (41%)	1	18	0
Second-degree SA block with Wenckebach phenomenon	6 (13%)	3	3	0
Junctional rhythm	23 (50%)	9	14	7
No bradydysrhythmia observed	9 (20%)	—	—	—

FIG. 4 Examples of episodes of third-degree sinoatrial block monitored by electrocardiogram. Upper strip: sinus rhythm, SA block, escape beat, sinus rhythm. Middle strip: atrial flutter, SA block, sinus bradycardia. Lower strip: atrial flutter, SA block, atrial flutter. Paper speed 50 mm/sec.

TABLE 5 Monitored sequence of electrocardiographic events in paroxysms of third-degree sinoatrial block

Monitored sequence, type of arrhythmia	No. of patients	No. of patients with cerebral symptoms during monitoring
Sinus rhythm, SA block, sinus rhythm	20	12
Atrial fibrillation, SA block, sinus rhythm	8	6
Atrial fibrillation, SA block, atrial fibrillation	2	2
Atrial flutter, SA block, atrial flutter	2	2
Atrial flutter, SA block, sinus rhythm	1	1
Atrial flutter, SA block, junctional rhythm	2*	1*
Atrial tachycardia, SA block, sinus rhythm	2	2
Atrial tachycardia, SA block, atrial tachycardia	1	0
Left atrial tachycardia, SA block, junctional rhythm	1	1
Junctional rhythm, SA block, junctional rhythm	2*	2*
Coronary sinus tachycardia, SA block, junctional rhythm	1	0
Total no. of patients	40	27

* One patient in each of these groups also had sinus rhythm, SA block, sinus rhythm, and is recorded twice, and both have been monitored by electrocardiogram during attacks.

followed by the establishment of an escape rhythm. Heart rate and QRS configuration of these rhythms appear in Fig. 4.

Table 5 shows the sequence of electrocardiographic events before and after the occurrence of third-degree sinoatrial block in the 40 patients. Examples of syncopal episodes monitored by electrocardiogram are shown in Fig. 4.

Drug treatment was attempted in 30 patients before pacemaker implantation. Digoxin often combined with quinidine and/or propranolol was used in 11 cases with tachydysrhythmias. No patient was improved, and an increase in the frequency of syncope was seen in 8 cases. Isoprenaline was administered against attacks of bradycardia and block in 14 cases and proved helpful in 6 cases, and atropine was given in 10 cases (with some beneficial effect in 5). However, no patient was relieved of syncope.

In 39 cases, a demand pacemaker was implanted. In all cases, the cerebral attacks disappeared. Twenty-four of these had had attacks of tachydysrhythmia before pacemaker implantation. In 3 cases these attacks disappeared after the pacemaker implantation. In the remaining 21 patients (Fig. 5) the implantation of a pacemaker allowed maximal drug treatment of tachydysrhythmias. The drug of choice was digoxin, often combined with quinidine and/or betablocking agents. With this treatment, 17 patients had nearly total relief from tachydysrhythmias, while the remaining 4 patients still had troublesome symptoms, caused by the frequent appearance of tachydysrhythmias.

Discussion

The present series confirms that syncope due to sinoatrial block usually is associated with other signs of sinus node dysfunction (Easley and Goldstein, 1971). Most patients (in this series 90 per cent) have frequent, more or less, sustained paroxysms of one or several different types of supraventricular dysrhythmias (Fig. 3). The many dysrhythmias, especially the attacks of tachydysrhythmia, often give rise to troublesome symptoms, such as attacks of palpitations, fatigue, and even congestion, and thus contribute to the discomfort of the patient, but they also make it easier for the clinician to recognize the cardiac origin of the highly unspecific cerebral symptoms.

Additional heart disease, primarily coronary artery

disease, is commonly but by no means constantly found. In this series, as in series of patients with the sick sinus node syndrome (Rasmussen, 1971; Rubinstein *et al.*, 1972), about one-fourth of all patients had sinus node dysfunction as the only manifestation of cardiac disease. Male-to-female ratio is about 1 to 1 and mean age for onset of symptoms is 63 years without significant difference in sex. These figures differ significantly from what should be expected, if coronary artery disease were the dominant aetiological factor, and they also differ significantly from those observed in patients with fainting due to atrioventricular block (Johansson, 1961; Zoob and Shirley Smith, 1963; Jensen *et al.*, 1973).

It seems, therefore, that other factors than coron-

FIG. 5 Sinus rhythm, AV junctional rhythm, and atrial fibrillation alternating with third-degree SA block. After pacemaker implantation the supraventricular tachydysrhythmia persists. During SA block, the ventricular rate is maintained by the pacemaker. The strips are continuous within each half of the picture. Paper speed 25 mm/sec.

FIG. 6 Heart rate and QRS configuration in escape rhythms. One patient had alternation between changed and unchanged QRS pattern in escape rhythm and is included twice.

any artery disease play an aetiological role in the development of sinoatrial block syncope, and that syncope due to sinoatrial and atrioventricular block has few, if any, aetiological factors in common.

The major electrophysiological deficiency which provokes syncope is not the sinoatrial block *per se*, but the failure of other pacemakers within the pacemaking system to function so that an adequate cardiac rate is not sustained. Intermittently, escape rhythms may develop. Unchanged QRS configuration, often with a slow rate, will then be the rule (see Fig. 6), whereas ventricular rhythms and even ventricular escape beats only rarely occur. This means that the depression of automaticity usually occurs to the same degree throughout the pacemaking system of the heart below the sinus node.

In a substantial number of patients, the episodes of asystole are linked to preceding episodes of tachycardia (Fig. 4 and 5). This observation may be explained by the depressive effect of rapid overdrive pacing on most pacemaking tissue (Pick, Langendorf, and Katz, 1951), an effect which recently has been shown to be more pronounced in patients with the sick sinus node syndrome (Narula, 1971; Rosen *et al.*, 1971).

Atrioventricular and interventricular conduction abnormalities were observed in the present study in only 20 per cent, whereas the incidence in previous studies of the sick sinus node syndrome varied between 20 and 60 per cent. Paroxysmal third-degree atrioventricular block has, to our knowledge, not been reported in patients with sick sinus node syndrome just as sinus node dysfunction seems to be infrequently found in patients with syncope due to paroxysmal third-degree atrioventricular block (Jensen *et al.*, 1973). Altogether this evidence

favours the assumption that Adams-Stokes disease caused by paroxysmal sinoatrial and by paroxysmal atrioventricular block are two fairly distinct pathological entities with little overlapping.

The cerebral symptoms, syncope as well as black-out episodes, may disable these patients, who are often only in middle-age, to a severe degree. They occur suddenly and unexpectedly, make the patients afraid to leave their homes, prevent them from taking part in social activities, and render them incapable of driving. Consequently, the first aim of any therapy should be to free the patients from the cerebral attacks, while prevention of tachycardia is of secondary importance. The best treatment is implantation of a demand pacemaker which will prevent the cerebral attacks and will also in a few cases favourably influence the tendency to tachycardia (Sandøe and Flensted-Jensen, 1969; Bayley, 1971). However, in most cases, the tachydysrhythmias will continue (Fig. 5). We – and other investigators – have used pacing through a catheter inserted in the right ventricle. Recently, atrial pacing on demand has been recommended as a means to preserve the haemodynamic benefits of synchronous atrial contraction (Samet *et al.*, 1965; Castillo *et al.*, 1971). The outcome of drug therapy (isoprenaline and/or atropine) has in this study and in earlier studies had no or only slight effect on the cerebral symptoms (Easley and Goldstein, 1971; Rasmussen, 1971).

The tendency to tachycardia may be influenced by preventive treatment with digoxin, quinidine, and/or beta-blocking agents after the insertion of a pacemaker, whereas initiation of such drug treatment before pacemaker implantation most often will lead to aggravation of the cerebral symptoms.

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